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Congenital superior sternal cleft repair using our modified Sally technique: A case report[☆]

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ABSTRACT

We report the case of an 8-month-old male neonate who underwent reconstructive surgery for a congenital superior sternal cleft to decrease the risk of commotio cordis. With regard to use of the Sally technique for the repair of a relatively wide cleft (4×4 cm), we were concerned about respiratory problems caused by compression following closure of the sternal halves. By closing the sternal halves imperfectly, elevation of intrapleural pressure can be avoided to some extent. By bridging the surplus resected cartilage from the lower sternum over the gap of the upper sternum, the heart is protected by more rigid material. We recommend our modified Sally technique because it is both flexible and effective for sternal cleft repair.

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Congenital superior sternal cleft is a rare anomaly caused by failed midline development and fusion of mesodermal lateral plates. We report on our surgical experience using modified Sally technique with a case of superior sternal cleft unassociated cardiac anomaly.

1. Case report

A male neonate delivered by elective cesarean section at 37 weeks of gestation with a birth weight of 2460 g presented with dyspnea and paradoxical breathing at birth, necessitating admission to the neonatal intensive care unit. He was the second child born of an uneventful pregnancy. His mother reported no exposure to medication or radiation during pregnancy, and there was no family history of congenital anomalies. Computed tomography (CT) revealed a superior U-shaped sternal cleft with widening between the medial ends of the clavicles. At 14 days of age, his respiratory status improved and he was discharged. Subsequently, he was referred to our hospital for repair of the superior sternal cleft. A pulsate heart was readily visible through the normal skin over the defect. He presented with unstable head and neck movements and

disordered functional motion of both upper limbs because both clavicles, which were attached to the sternocleidomastoid and trapezius, were not fixed to the manubrium. No infantile hemangioma of the head or umbilical raphe was detected. In addition, he presented with callosal hypoplasia and cutaneous syndactyly between the third and fourth fingers. His laboratory test results were within normal limits and his respiratory status was stable. No cardiac anomaly was confirmed on echocardiography. The brain surgeon followed the course of callosal hypoplasia. The patient steadily gained weight, but at the age of 5 months, unstable head and neck and disordered upper limbs movements persisted. We were concerned about the risk of commotio cordis because of increasing activity with no hard support from the anterior thorax; we performed the first surgery at the age of 8 months (body weight, 6.4 kg). Pre-operative three-dimensional CT showed that the cleft was 4.7 cm wide at the level of the sternoclavicular junction (Figs. 1–5).

1.1. Surgical method

A vertical midline incision was made from superior end of the defect to the xiphoid process, and a skin flap was laterally raised to expose the sternal cleft. The pectoralis major muscles were raised from their corresponding medial chondral insertions. When the sternal tables were encountered, the superficial and deep surfaces were freed laterally to the intercostal spaces. Then, the sternal halves were fused at the caudal end to the cartilaginous xiphoid. This portion was excised by dividing the sternal attachment parallel

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Fig. 1. Preoperative period.

to the defect. The perichondrium was elevated anteriorly and posteriorly on each sternal half to expose the underlying cartilage. Then posterior perichondrium of the sternum was approximated upward with several 2-0 absorbable sutures from the lower end. To ensure that there were no complications, we monitored his cardiac and respiratory functions for a while before the sutures were tied. Central venous pressure increased in proportion with suture tension. To prevent a deterioration in hemodynamics and respiratory status, the manubrium was not sutured. The surplus resected cartilage from the lower sternum was used to bridge the gap of the upper sternum. Following sternal reconstruction, the previously raised muscles were sutured back to cover the cartilage grafts and the incision was closed.

2. Results

The surgical duration was 4 h and 11 min, and the blood loss volume was 40 cc. Blood transfusion was not required. After a 24 h observation period in the pediatric intensive care unit, the patient steadily recovered and was discharged on postoperative day 9. At postoperative month 6, his functional and cosmetic outcomes were good and direct cardiac pulsation was no longer visible. His head and neck became stable and function of both upper limbs improved. CT at postoperative month 6 confirmed survival of the bridged cartilage grafts and fusion of the sternal halves. The interclavicular gap decreased from 4.7 cm to 2.7 cm.

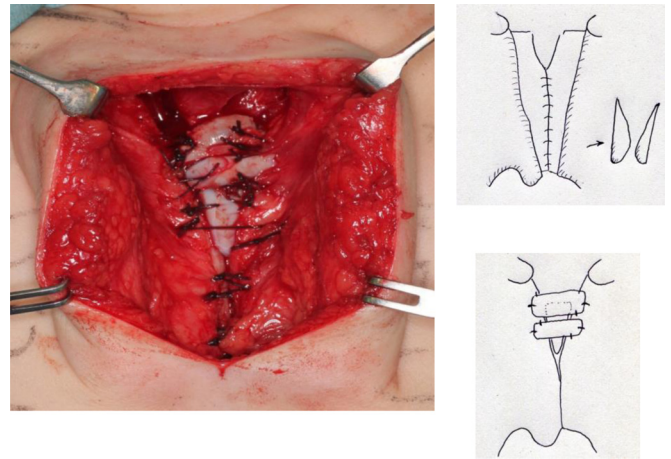


Fig. 3. Intraoperative finding of cartilaginous grafts bridging the gap.

3. Discussion

During the sixth week of gestation, paired mesenchymal bars form lateral to the midline and parallel to each other. These sternal bands then migrate toward the midline and fuse in a cephalocaudal direction to form the sternal plate. Sternal clefts result from fusion failure of the sternal bands by the eighth week of gestation [1]. Although the etiology of sternal clefts remains unclear, alcohol intake, methylcobalamin deficiency, and disruption of *Hoxb-4* gene function have been inconclusively associated with defects in mice [2]. Sternal clefts were first reported in 1740 in London, the first attempt at surgical correction was reported by Lannelongue in 1888, and the first successful repair of a sternal anomaly was performed by Burton in 1943 [3].

Hersh [4] suggested a sternal cleft classification system consisting of four categories: (I) cleft sternum without associated anomalies, (II) cleft sternum with vascular dysplasia, (III) true ectopia cordis, and (IV) Cantrell's pentalogy (modified Ravitch classification). Acastello et al. [5] reported that sternal clefts accounted for 0.15% of all chest wall malformations and can be complete or incomplete. The most common form of sternal cleft is the partial superior type, accounting for 67% of all patients, followed by the complete form (19.5%) and the partial inferior type (11%) [6]. Sternal clefts are most commonly diagnosed in the neonatal period and are often associated with cardiac anomalies; vascular anomalies; supraumbilical raphe; posterior fossae of the brain, arterial anomalies, cardiac anomalies, and eye anomalies

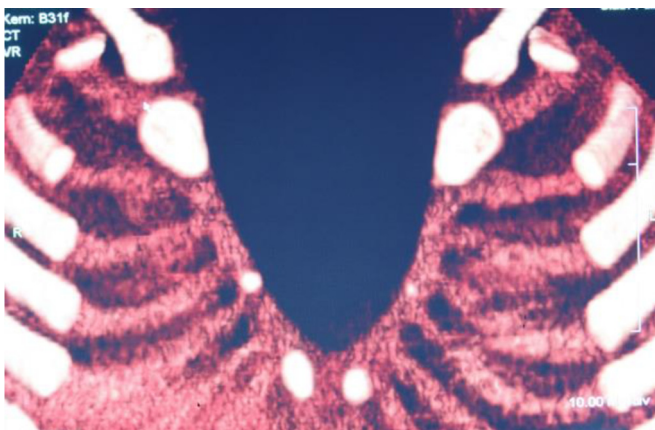


Fig. 2. A preoperative computed tomography image (4 × 4 cm defect in the upper sternum).

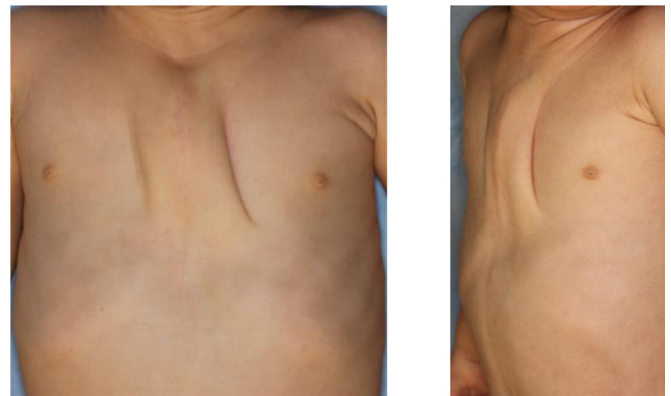


Fig. 4. Postoperative month 6.



Fig. 5. A computed tomography image obtained at postoperative month 6.

(PHACE) syndrome; and Cantrell's syndrome. In the literature, 73 cases of superior sternal cleft have been reported between 1947 and 2011 [3–38]. Among these reports, superior sternal clefts were more frequent in females than in males [40(55%) vs 21(29%)]; 12(16%) cases were of unknown gender. In Japan, nine patients (3 males, and 6 females) with superior sternal clefts were reported between 1930 and 2013, indicating a female predominance in accordance with the findings of other reports (Table 1). Of these nine patients, six had facial venous malformations, and one had supraumbilical raphe. The lack of bony protection increases the susceptibility the heart and great vessels to trauma, dyspnea, decreased lung aeration, and cough reflex, which could lead to an increase in the incidence of respiratory infection. Patients with large sternal cleft defects have a greater risk of fatal arrhythmia due to commotio cordis. Therefore, surgical repair of these defects is recommended during the neonatal period, when a compliant thorax allows direct approximation of the sternal halves.

The main goals of sternal cleft repair are to restore bony protection to the mediastinal viscera with respiration and to eliminate visible deformity.

3.1. Surgical timing

Maier et al. [7] reported that during the neonatal period, the chest wall is more flexible with lower resistance, but after the age of 4 weeks, the chest wall loses its flexibility and it becomes difficult to

directly suture the sternal halves. Therefore, chest wall reconstruction is highly recommended after the age of 4–6 weeks. We agree that surgery for sternal cleft repair should be performed during the neonatal period, but potential complications should be considered to maintain hemodynamics and respiratory status. In patients with cardiac anomalies in particular, cardiac surgery is preferentially recommended, and chest wall reconstruction should be performed after hemodynamics have improved. With regard to the timing for surgery, the decision should take into consideration not only the ease of surgery but also the status of the patient and potential risks. In our case, surgery was performed at the age of 8 months because of potential complications and because the chest wall was relatively flexible. The sternal halves were almost approximated, with the exception of the manubrium.

3.2. Chest wall reconstruction

Many techniques for chest wall reconstruction have been reported, which include primary closure, cartilage resection (Sally technique), sliding chondrotomy (Sabiston technique), cartilage graft bridging, and prosthetic closure using titanium plates, among others. Primary closure is performed by direct approximation of both sternal halves. The Sally technique [8] incorporates resection of cartilaginous xiphoid and approximation of both sternal halves. Sabiston [9] described a sliding chondrotomy technique in which oblique incisions through the costal cartilage are made to increase the length of the cartilage. Cartilage graft bridging is a technique by which the gap of the sternal cleft is bridged using costal cartilage. Repair of the defect with different autologous grafts (i.e., costal cartilage [10], ribs, parietal bone of the cranium, and tibia) has been reported. We do not prefer to employ prosthetic materials in sternal cleft reconstruction, because of the risk of infection and the inability of these inert materials to remodel along with patient growth. The advantages and disadvantages of each technique are presented in Table 2. Of the reported 73 cases patients, 14 (19%) achieved primary closure, 17 underwent sliding chondrotomy (Sabiston technique), 19 received cartilage or bone grafts, six received a prosthesis, two underwent cartilage resection (Sally technique), four underwent surgery with other techniques, eight underwent treatments with no available data, and three did not undergo surgery. Primary closure via cartilage/bone grafting and chondrotomy (Sabiston technique) were most preferable. In Japan, eight (88.9%) of nine patients with superior sternal clefts underwent chest wall reconstruction: three underwent primary closure, two patients underwent chondrotomy (Sabiston technique), two

Table 1
Patients and methods.

Number	Gender	Age ^a	Date ^b	Technique
1	Male	16 yr.	1930	—
2	Female	1 mo.	1973	Unknown
3	Female	16 da.	1978	Sliding chondrotomies (Sabiston)
4	Female	18 da.	1979	Primary closure alone
5	Female	41 da.	1991	Primary closure alone
6	Female	48 da.	1995	Sliding chondrotomies (Sabiston)
7	Female	81 da.	1995	Cartilage resection (Sally)
8	Male	89 da.	2000	Primary closure alone
9	Male	8 mo.	2013	Cartilage resection (Sally) + Cartilage graft

^a Age: operation age.

^b Date: reported date.

Table 2
Advantages and disadvantages.

	Advantage	Disadvantage
Primary closure	Simple technique	Increase intrathoracic pressure, short range of adaptation age
Cartilage resection (Sally technique)	Simple technique	Increase intrathoracic pressure, longer range of adaptation age
Cartilage graft bridging	Prevent increasing intrathoracic pressure	Donor expense, widening between cartilage graft associated growth
Sliding chondrotomy (Sabiston technique)	Prevent increasing intrathoracic pressure	Cumbersome technique, hemorrhagic risk
Prosthetic closure	Prevent increasing intrathoracic pressure	Cumbersome technique, infection risk

underwent cartilage resection (Sally technique), and one underwent treatment with no available data. Primary closure was most preferable. During the neonatal period, primary closure is recommended because the chest wall is relatively flexible. In cases of delayed consultation or timing for various reasons, as in our patient, the chest becomes more rigid and approximation of both sternal halves more difficult. The Sally technique is a relatively simple and less invasive method. When both sternal halves are approximated, intrapleural pressure is elevated. We found that elevated intrapleural pressure can be avoided to some extent by imperfectly closing the gap. The surplus resected cartilage from lower sternum was used to bridge the gap of the upper sternum. Survival of the bridged cartilage graft was confirmed by CT at postoperative month 6. For our patient, we supported the gap using only surplus resected cartilage, although costal cartilage graft transplantation is required in patients with large gaps. We recommend this method because it is effective, less invasive, requires less time, and employs a simple procedure to compensate for the gap. If cardiac embarrassment is imminent at any stage, repair must be abandoned for more extensive repair using an autologous free graft.

4. Conclusions

In conclusion, there are various surgical techniques for chest wall reconstruction. Our modified Sally technique is flexible and effective. Considering the risk of complications because of changes in chest wall morphology accompanied by growth, long-term follow-up is necessary in such patients.

Conflict of interest

The authors have no conflicts of interest to disclose.

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